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Synchronous tumors: clear cell carcinoma of pancreas and gastrointestinal stromal tumor of stomach

Senkron tümörler: pankreas berrak hücreli adenokarsinomu ve mide gastrointestinal stromal tümör

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Abstract

Although clear cell carcinoma is a common variant of carcinoma of kidney, ovary, thyroid and lung, this tumor rarely originates from the pancreas. On the other hand, gastrointestinal stromal tumors represent the most usual mesenchymal tumors affecting the gastrointestinal tract, and are mostly located in the stomach and small bowel.

In this paper, we report a clear cell variant of pancreas adenocarcinoma combined with gastrointestinal stromal tumor of the stomach in a 76-year-old woman who presented with right upper quadrant pain, jaundice, nausea and vomiting.

Synchronous appearance of gastrointestinal stromal tumors and pancreatic adenocarcinoma has been rarely reported. To our knowledge, this is the first report about clear cell variant of pancreas adenocarcinoma combined with gastric gastrointestinal stromal tumor in the literature.

Keywords: Pancreas Adenocarcinoma; Clear Cell Carcinoma of Pancreas; Gastrointestinal Stromal Tumor.

Öz

Berrak hücreli karsinom, böbrek, over, akciğer ve tiroid karsinomlarının görece sık görülen bir varyantı olmasına karşın, bu tümör nadiren pankreastan kaynaklanmaktadır. Öte yandan, gastrointestinal stromal tümörler, gastointestinal sistemi etkileyen en sık mezenkimal tümörlerdir ve sıklıkla mide ve ince barsaklarda yerleşirler.

Bu yazıda, sağ üst kadran ağrısı, sarılık, bulantı ve kusma ile başvuran 76 yaşındaki kadın hastada mide yerleşimli gastrointestinal stromal tümör ile birlikte olan pankreas berrak hücreli karsinom olgusu sunuyoruz.

Senkron gastrointestinal stromal tümör ve pankreatik adenokarsinom vakaları literatürde nadiren bildirilmiştir. Bildiğimiz kadarıyla bu olgu, mide gastrointestinal stromal tümör ile pankreas adenokarsinomu berrak hücreli varyantı birlikteliği ile ilgili literatürdeki ilk vakadır.

Anahtar Kelimeler: Berrak Hücreli Adenokarsinom; Gastrointestinal Stromal Tümör; Pankreas Kanseri.

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INTRODUCTION

Pancreatic ductal adenocarcinoma (PDA) is one of the most common malignancies in human, with a poor prognosis. Over 90 of carcinomas diagnosed in the pancreas are classified as ductal in origin. According to the WHO classification, clear cell carcinoma of the pancreas is very rare and is classified as a "miscellaneous" carcinoma (1). Indeed, clear cell carcinoma is a common variant of carcinoma of kidney, ovary, thyroid and lung, but this tumor rarely originates from the pancreas, and is characteristically rich in glycogen and poor in mucin (2, 3).

Gastrointestinal stromal tumors (GISTs) are uncommon mesenchymal neoplasms of the gastrointestinal tract, accounting for 0.1-3% of all gastrointestinal malignancies (4). These tumors are arising from precursor interstitial cells of Cajal, and are mostly located in the stomach and small intestine.

The coexistence of adenocarcinoma and GIST is uncommon, and GIST is often detected incidentally during surgery or in pathological evaluation. Here, we present and discuss a rare case of synchronous gastric GIST and clear cell carcinoma of pancreas in an elderly female patient.

CASE REPORT

A 76-year-old female was admitted to our unit with abdominal pain, jaundice, nausea and vomiting. Physical examination was significant for epigastric and right upper quadrant tenderness to palpation but the patient had no palpable abdominal masses. She had no significant medical history or prior history of cancer. Her serum CA 19-9 level was elevated (>700 U/ml), but other routine laboratory parameters were all within normal limits. Abdominal computed tomography showed a pancreatic mass, 20 mm in size, with dilated duct of Wirsung. Magnetic resonance imaging revealed a mass at the pancreatic head-corpus junction, 24×20 mm in size, with milimetric cystic areas in the centre of the lesion. Main pancreatic duct was significantly dilated distal to the lesion (max. 10 mm). Portal vein and superior mesenteric artery were patent (Figures 1A, B).

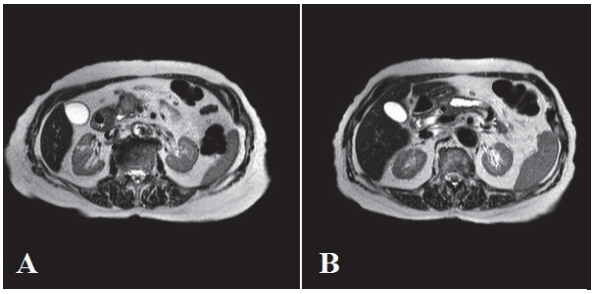

Figure 1. Axial T2 weighted image shows (A) mild hyperintense mass of the pancreas and (B) dilatation of the pancreatic duct and atrophy of the distal part of the pancreas.

There was no evidence of other primary or secondary tumors outside the pancreas. Pancreaticoduodenectomy was performed for the tumor. On histopathological examination, an adenocarcinoma, 3.5×3×2 cm in size, which was composed of glycogen-rich and mucin-poor tumoral cells was determined in the head of pancreas, defined as clear cell carcinoma (Figure 2A). Three of 30 lymph nodes detected in peripancreatic region were metastatic. In addition, a subserosal nodular lesion, 10 mm in diameter, was incidentally found in the wall of stomach. Immunohistochemical staining for C-kit and CD34 was positive, while that for SMA, desmin and S-100 was negative for gastric tumor, called as GIST (Figure 2B).

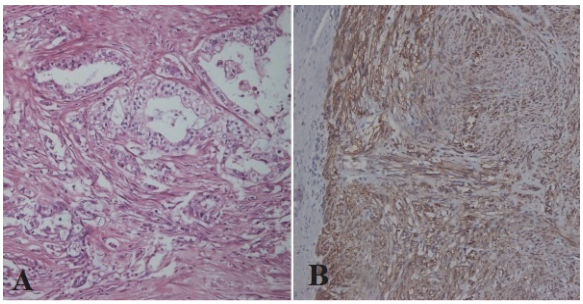

Figure 2. (A) Infiltration of neoplastic glands composed of atypical cells with clear cytoplasm in desmoplastic stroma (H&E X200), (B) Cytoplasmic immunoreactivity of CD-117 (c-KIT) in the gastrointestinal stromal tumor cells (CD-117 X200)

DISCUSSION

GISTs and adenocarcinoma are distinct neoplasms originating from different cell layers, and the coexistence of these tumors is an uncommon condition. It is not clear whether this is a simple incidental coexistence or the two lesions are connected by a causal relationship. Gene mutations or another reason that has not been known yet, may underlie this rare situation. GIST, as a synchronous tumor, has often been detected incidentally during the operation or in pathological evaluation. In our case, the preoperative imaging methods showed only a tumoral lesion located at the head of pancreas. There was no evidence of another abdominal mass, and the GIST was detected in the pathological examination of the resected stomach. In fact, many cases of synchronous or asynchronous GIST with other gastrointestinal tumors have been reported to date, and an increased risk of additional cancers were found among patients with GISTs (5). However, the coexistence of pancreatic cancer and GIST of stomach is extremely rare. Tavares et al reported a pancreatic neuroendocrine tumor combined with GIST of the stomach which was found during surgery (6). Similarly, a case of adenosquamous carcinoma of pancreas and synchronous gastric GIST detected in surgical spesimen, was reported by Dasanu et al (7). In a study by Wronski et al, it was determined that the synchronous occurrence of GISTs and other gastrointestinal malignancies is more common than it has been considered (8). They found synchronous occurrence of GISTs with another

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gastrointestinal malignancy with a proportion of 14% in their work. The authors also suggested that the development of GIST and other neoplasms may involve the same carcinogenic agents. Similarly, in a study published by Liu et al, 17.4% of gastric GISTs were found to be associated with other gastrointestinal epithelial malignant tumors (9). They also reported that a total of 90.7% incidental GIST lesions occurred in stomach, and the most common sites were the gastric fundus and body. In addition, most of the GISTs were small in size with a low mitotic activity and very low risk of malignancy. Therefore, the majority of cases do not require any additional surgical intervention. In our case, GIST was incidentally found in the resected distal stomach which was the routine part of Whipple prodecure, and no another surgical intervention was needed due to the small size and low grade of the tumor.

In conclusion, the coexistence of pancreatic carcinoma and GIST of stomach has been reported in a small number of case; however, synchronous gastric GIST with clear cell variant of pancreas adenocarcinoma has not yet been reported. The underlying pathogenesis of concurrence of GIST and other tumors is still unclear, and further studies are required to clarify the molecular and genetic mechanisms of this interesting condition.

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